Successful Removal of Multiple Cerebellar Hemangioblastomas. Case report

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Successful Removal of Multiple Cerebellar Hemangioblastomas. Case report

by

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Summary

The unusual case of a 15 year old girl with four anatomically discrete hemangioblastomas is reported. The first and second tumors were located in each cerebellar hemisphere and the third tumor in the area postrema. The fourth tumor was huge and dumbbell-shaped; the massive cystic part of which located in the central portion of the cerebellum and the solid part herniated upwards into the tectal region through the incisura tentorii.

Total removal of all tumors was successfully accomplished in stages at three separate operations. Histological examination revealed precisely the same morphology in all the hemangioblastomas.

Because of the depth and delicate location of the last tumor, extirpation via the suboccipital approach alone was deemed too risky, accordingly a combined left temporo-occipital suboccipital craniotomy was employed.

Hemangioblastomas of the central nervous system are histologically benign vascular neoplasms which occur predominantly in the cerebellum. An excellent prognosis is expected if they are totally removed. The staged total excision of multiple cerebellar hemangioblastomas accomplished at three separate operations is reported.

Case Report

A 15 year old right-handed girl was admitted to the Kobe Municipal Central Hospital on March 1, 1972 with complaints of headache, nausea, vomiting and nuchal pain over a month prior to admission. The family history revealed that an uncle had died of a cerebellar cystic tumor at a university hospital.

Examination: The general physical findings were unremarkable. Neurological

Key words: multiple cerebellar hemangioblastoma, transtentorial upward herniation, combined craniotomy.

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examination revealed an alert and oriented girl with minimal nuchal rigidity and symmetrically hyperactive deep tendon reflexes. Neither pathological reflexes nor nystagmus could be detected. The finger-nose test was clumsy on the left side. A careful examination of the optic fundi failed to reveal abnormal vessels or papilledema.

The hematocrit was 43% and all routine blood chemistries were normal. A lumbar spinal tap yielded crystal clear C.S.F. under a pressure of more than 300 mm in water with protein of 330mg/dl, sugar of 58mg/dl and chloride of 122mEq/l.

Skull and chest films were normal. A right carotid angiogram showed changes suggesting ventricular enlargement. A right retrograde brachial vertebral angiogram revealed 4 tumor stains in the posterior cranial fossa. (Figs. 1 and 2) A Conray ventriculogram disclosed kinking and obstruction of the aqueduct. (Fig. 3)

1 st operation: On March 23, under general endotracheal anesthesia with the patient in the sitting position, a suboccipital craniectomy with first cervical laminectomy was performed and a cystic tumor with a mural nodule (the first tumor) in the right cerebellar hemisphere was removed. The histological diagnosis was hemangioblasto ma. (Fig. 4)

2 nd operation: On April 12, the previous incision was reopened and 2 solid tumors, one in the left cerebellar hemisphere (the second tumor) and the other in the area postrema (the third tumor) were totally removed.

Her postoperative course was initially uneventful until eleven weeks after the second operation, when she developed occipital pain and urinary retention. Vertebral angiography showed a huge tumor stain in the tectal region with changes suggesting transtentorial upward herniation. (Fig. 5) A third operation was therefore indicated.

3 rd operation: The last tumor was so deep and delicately placed that total removal was thought to be impossible via a suboccipital approach only. Therefore, a left temporo-occipital and suboccipital combined craniotomy was undertaken on July 19. A huge, dumbbell-shaped cystic tumor was found in the central portion of the cerebellum. It protruded upwards through the incisura tentorii where there was the solid part of the tumor with a loose attachment to the dorsal surface of the midbrain (Fig. 7) Through the simultaneous subtemporal and suboccipital approach the solid tectal part of the tumor was excised completely but with considerable difficulty.

Histological examination of all 4 discrete tumors revealed the same well-differentiated morphology; hemangioblastoma. The nature, size, location and appearance of the lesions are shown in Table 1.

Postoperative course: The postoperative course was uneventful. However, right homonymous hemianopia, diplopia and slight incoordination of gait have been noted. Postoperative vertebral angiography revealed neither tumor stain nor abnormal displacement of major vessels.

Discussion

Hemangioblastomas are histologically benign vascular neoplasms of the central nervous system which usually arise in the cerebellum. RUSSELL[5] states that in the large series reported from neurosurgical clinics the hemangioblastomas form from 1.1 to 2.4% of all intracranial tumors. In the adult, the incidence amongst primary

Table 1.

	Location	Gross appearance	Size	Nature
1st tumor	Right cerebellar hemisphere	Cystic		A thin-walled cyst, with clear amber fluid
		A mural nodule	7 mm	Well demarcated, spherical and reddish
2nd tumor	Left cerebellar hemisphere	Solid	4 mm	Well demarcated, spherical and reddish
3rd tumor	Area postrema	Solid	20×20×20mm	Well demarcated, non-encapsulated, spherical, reddish and highly vascular
4th tumor	Central portion of cerebellum	Cystic		A thin-walled cyst, with clear amber fluid
	Tectal region	Solid	15×17×22mm	Well demarcated, non-encapsulated, spherical, reddish and highly vascular

tumors of the posterior fossa is 7.3%. Males are more frequently affected than females[4]. It is said that this neoplasm is not manifest in early life, and in most series the average age is 40 years[3]. In the present case, however, the lesions were clinically manifest at the age of 15 years.

In the cerebellum, the hemangioblastomas occupy a variety of sites; a paramedian hemispheric position is especially favored, but lateral lobes and vermis are also commonly affected. The tumor also arises in the medulla oblongata, particularly at the area postrema. Though usually solitary, more than one tumor may arise in the posterior cranial fossa[5].

STEIN et al[7]. have described multiple hemangioblastomas in the cerebellum which they interpreted as separate growths rather than recurrence of an incompletely resected tumor. Similarly, Cramer and Kimsey[1] from experience with 53 cerebellar hemangioblastoma cases treated by multiple operations concluded that this entity is not infrequently multicentric in this region.

Vertebral angiograms are known to be valuable in demonstrating the presence of multiple hemangioblastomas[4]. Vertebral angiograms can reveal not only location but also arterial supply and venous drainage of the tumors permitting control of bleeding and total removal of the lesions[2]. As described above, this patient had 4 discrete tumors which were visualized by vertebral angiography and verified at operation.

Supratentorial hemangioblastoma is extremely rare[3],[5]. Angiography seemed to indicate that the solid part of the fourth tumor was supratentorial in the tectal region. At operation, however, it became clear that it was not a true supratentorial lesion but that the solid part of the tumor had protruded upwards through the tentorial incisura from the posterior fossa to occupy the tectal region by the mechanism of so-called transtentorial upward herniation.

Hemangioblastoma is also known to be associated with erythrocytosis with

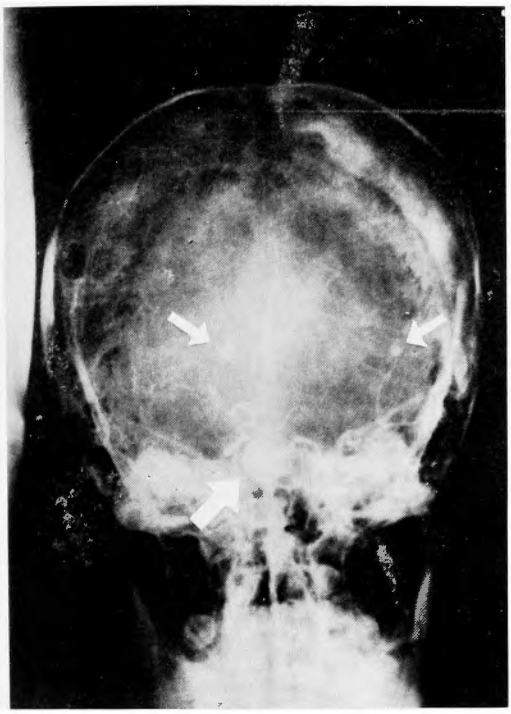
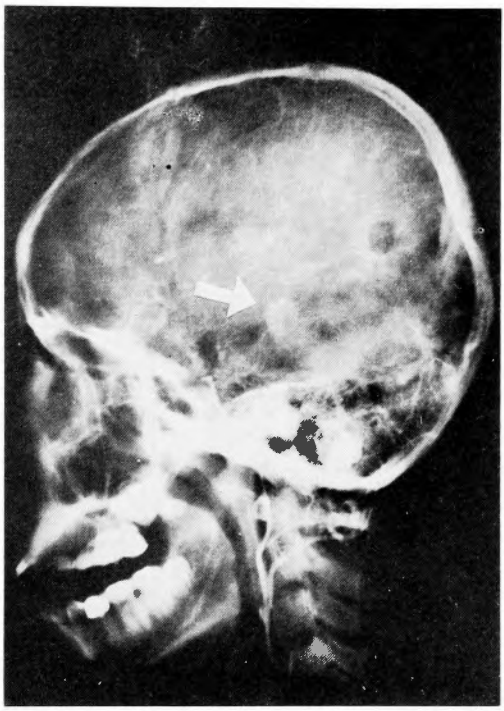


Fig. 1.



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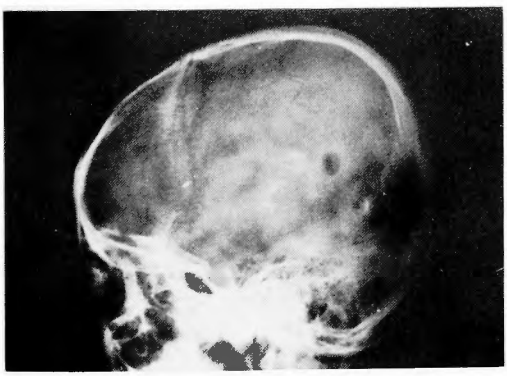

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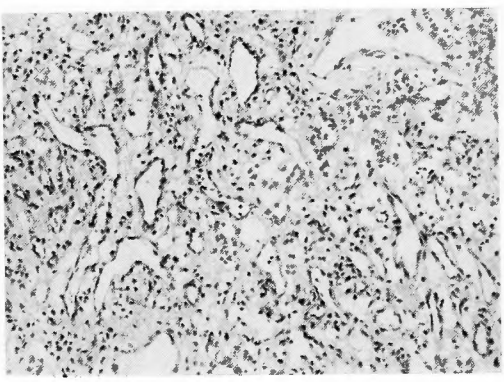

Fig. 4.

Fig. 1. Vertebral angiogram (A-P view), showing 3 areas of tumor stain in the posterior cranial fossa.

The 1st and 2nd tumors in each cerebellar hemisphere are indicated by thin arrows, and the 3rd tumor in the area postrema is indicated by a thick arrow.

Fig. 2. Vertebral angiogram (Lateral view), showing 4 tumor stains ; the 4th tumor in the tectal region (indicated by an arrow).

Fig. 3. Conray ventriculogram, showing the kinking and obstruction of aqueduct.

Fig. 4. Photomicrograph shows fine mesh of the blood-spaces and channels of the capillary structure. Histological diagnosis is hemangioblastoma, H. and E., 100×

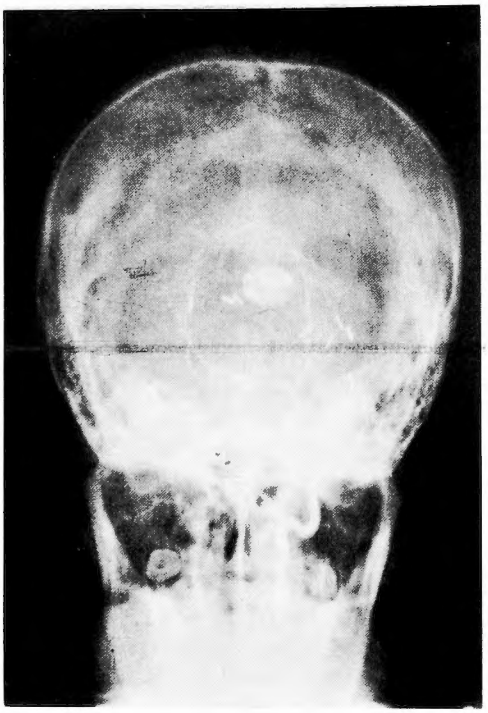

Fig. 5.

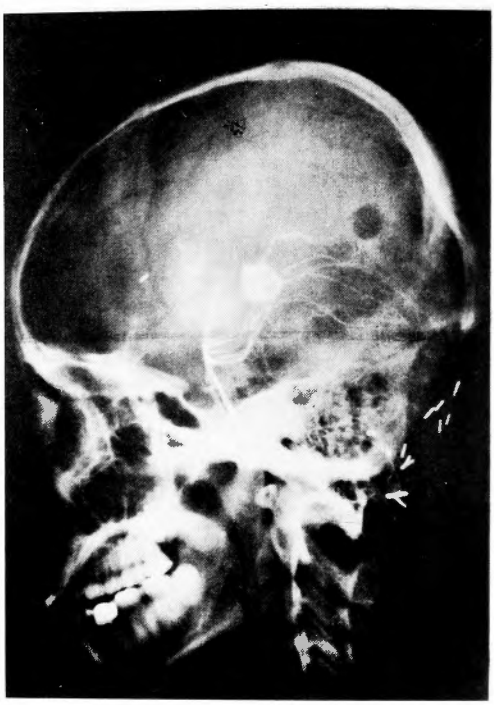

Fig. 6.

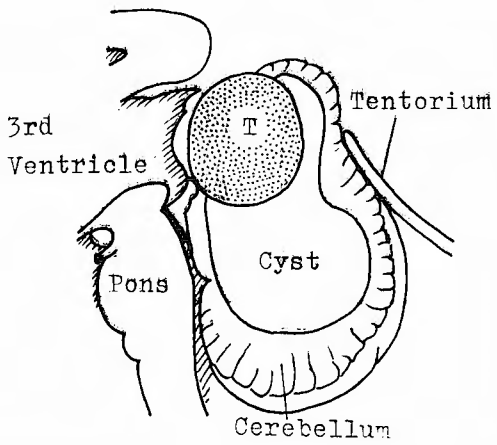

Fig. 7.

Fig. 5, and 6. Vertebral angiograms (A-P and lateral views), disclosing a huge tumor stain in the tectal region with the changes suggesting the transtentorial upward herniation.

Fig. 7. Schema of the 4th tumor. The solid part of the 4th tumor protruded upwards through the tentorial incisura from the posterior fossa to occupy the tectal region by mechanism of so-called transtentorial upward herniation.

abnormally high levels of hemoglobin[1),6),8)], but this was not the case in our patient.

The literature documents the excellent prognosis of hemangioblastoma following successful complete surgical removal. Therefore, total excision is recommended as the treatment of this benign tumor. Although total removal becomes much more difficult in cases where the tumors are deeply situated and/or multiple, a vigorous approach is required to effect a cure. Great effort and staged procedures may be necessary to achieve a total excision as in this case where 4 discrete tumors were removed in 3 operations.

References

1) Cramer, F., Kimsey, W.: The cerebellar hemangioblastomas: review of fifty-three cases, with special reference to cerebellar cysts and the association of polycythemia. Arch. Neurol. Psychiat., **67**: 237-252, 1952.
2) Garcia-Bengochea, F., Berk, M.: The so-called solid hemangioblastomas of the cerebellum and vertebral angiography. J. Neurosurg., **22**: 35-39, 1965.
3) Ishwar, S., Taniguchi, R. M., Vogel, F. S.: Multiple supratentorial hemangioblastomas: Case study and ultrastructural characteristics. J. Neurosurg., **35**: 396-405, 1971.
4) Mondkar, V. P., Mckissock, W., Ross Russell, R. W.: Cerebellar haemangioblastomas. Brit. J. Surg., **54**: 45-49, 1967.
5) Russell, D. S., Rubinstein, L. J.: Pathology of tumours of the nervous system. E. Arnold, London, pp. 72-76, 1963.
6) Silver, M. L., Hennigar, G.: Cerebellar hemangioma (hemangioblastoma): A clinicopathological review of 40 cases. J. Neurosurg, **9**: 484-494, 1952.
7) Stein, A. A, Schilp, A. O., Whitfield, R. D.: The histogenesis of hemangioblastoma of the brain: A review of twenty-one cases. J. Neurosurg., **17**: 751-761, 1960
8) Ward, A. A., Jr., Foltz, E. L., Knopp, L. M.: "Polycythemia" associated with cerebellar hemaogioblastoma. J. Neurosurg., **13**: 248-258, 1956.

和文抄録

多発性血管芽腫の1治験例

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伴　貞彦，尾形誠宏

解剖学的にそれぞれ独立した，4箇の多発性血管芽腫を有する稀有な1症例を経験した．

患者は15才の女子で，第1と第2腫瘍は左右小脳半球に，第3腫瘍は Area postrema にあった．第4腫瘍は小脳の中心部に位置し，一部は天幕切痕を通して上向性に天幕ヘルニアを起こしていた．これら4箇の腫瘍を3回にわたる手術で全摘した．なお，第4の腫瘍は巨大で，亜鈴状を呈し，深部でデリケートな位置にあったため，左側頭後頭開頭術と後頭下開頭術を組合わせた Supra- and infra-tentorial approach により全摘に成功した．病理組織診断で，4箇の腫瘍は等しく血管芽腫であった．